BRIEF ARTICLES

Sterile Neutrophilic Folliculitis with Vasculopathy in a Young Male Patient with Infective Endocarditis

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ABSTRACT

Sterile neutrophilic folliculitis with vasculopathy is a rare entity. Histopathologically it is characterized by neutrophilic or suppurative and granulomatous folliculitis accompanied by a folliculocentric vasculopathy.[1] It is a cutaneous manifestation of a systemic illness or infectious triggers. Variations in clinical presentation are independent to the underlying medical condition.[1] Prompt identification may uncover an underlying systemic disease. The rarity of this clinical entity has led to a paucity of evidence regarding its etiology, diagnosis, and treatment recommendations. Herein, we present a case of sterile neutrophilic folliculitis with vasculopathy in a 34-year-old male hospitalized for infective endocarditis.

CASE REPORT

A 34-year-old male with a past medical history of hepatitis C and intravenous (IV) drug abuse was admitted to the hospital with methicillin sensitive *Staphylococcus aureus* (MSSA) infective endocarditis complicated by septic pulmonary emboli, cervical osteomyelitis with soft tissue extension, bacteremia and acute kidney injury. On hospital day four, the patient abruptly developed a folliculitis-like rash on bilateral upper extremities for which dermatology was consulted. He denied any associated symptoms including itching, burning or pain. Prior to consultation, he was treated with vancomycin, cefazolin, linezolid, and dialysis.

On physical examination, the patient had pink to purple papules on the bilateral distal upper extremities (Figure 1), some of which were eroded. Distal lower extremities revealed scattered, non-blanching, bright pink macules. On re-examination three days later, the lesions on the bilateral upper extremities had evolved into larger, inflamed, firm, pink, centrally umbilicated, semi-translucent papules (Figure 2), with few newly developed scattered similarly appearing lesions on the bilateral lower extremities. The previous macules on lower extremities persisted and were unchanged. Seven days after initial presentation, the lesions began to spontaneously involute, leaving pink, crusted papules and macules without scars. The eruption continued to fade over the course of two weeks despite

continued fevers and persistently positive blood cultures.

An extensive laboratory workup revealed a normal complete blood count and complete metabolic panel with exception of hemoglobin 9.9 g/dL (12.5-17), and creatinine 2.91 mg/dL (0.53-1.30). Hematoxylin and eosin stains of punch biopsies obtained on initial presentation and three days later demonstrated folliculocentric neutrophilic dermatitis with perifollicular vasculitic changes (Figure 3). A tissue culture from the second biopsy was negative for bacteria, deep fungal infection, and acid-fast bacilli. The histopathologic findings and negative cultures led to the final diagnosis of sterile neutrophilic folliculitis with perifollicular vasculopathy.

Figure 1. Pink to purple papules with some erosion on the bilateral distal upper extremities.

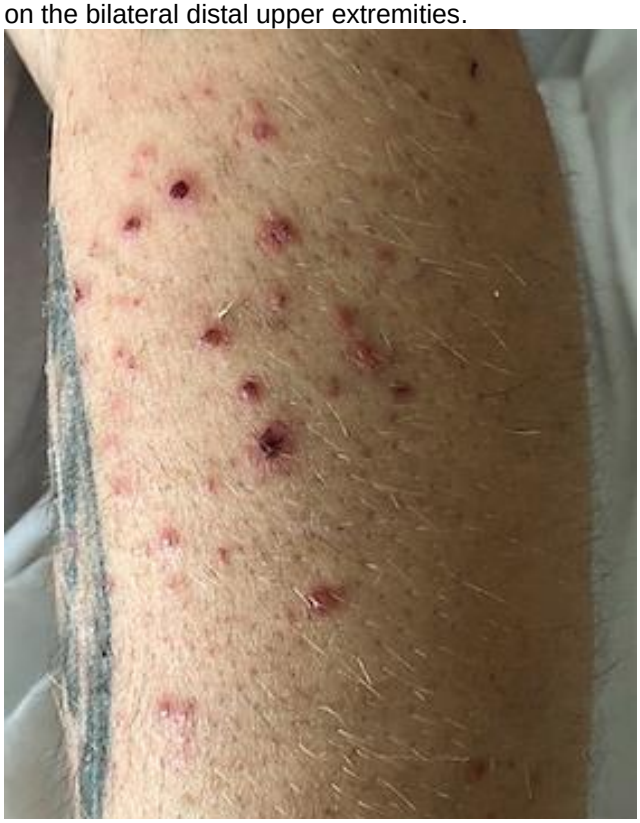


Figure 2. Inflamed, firm, pink, centrally umbilicated, semi-translucent papules on bilateral upper extremities.

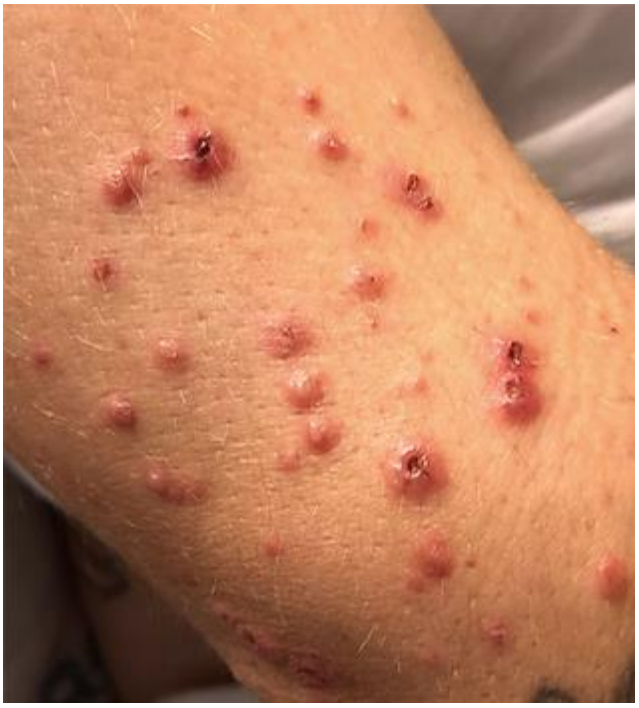
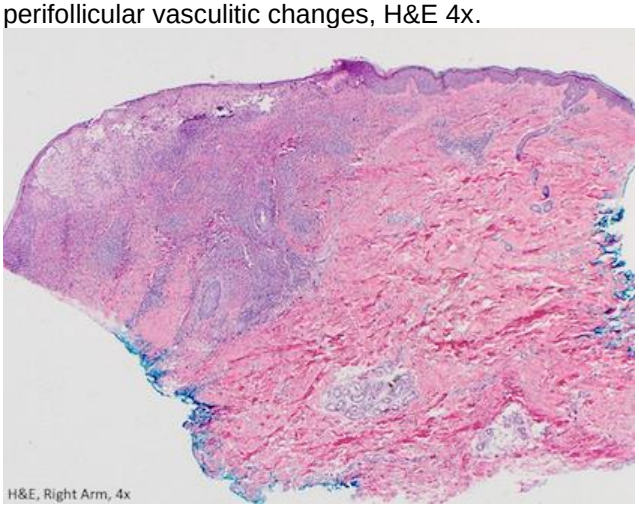


Figure 3. Folliculocentric neutrophilic dermatitis with perifollicular vasculitic changes, H&E 4x.



DISCUSSION

Sterile neutrophilic folliculitis with vasculopathy is a distinctive cutaneous pattern first described by Magro and Crowsen in 1998.[1,2] This rare entity is characterized by a constellation of

histopathological findings, which consist of neutrophilic or suppurative and granulomatous folliculitis in addition to a Sweet's-like vascular reaction or leukocytoclastic vasculitis. It is believed to be a reaction pattern to underlying systemic diseases such as Behcet's disease, reactive arthritis, inflammatory bowel disease, hepatitis B, various connective tissue diseases, and distant extracutaneous infections.[1,3]

Though it is speculative, the association between sterile neutrophilic folliculitis with vasculopathy and an underlying systemic disease the exact etiology remains unclear. A prominent theory proposes that it is caused by an aberrant humoral or cell-mediated immune response to various triggers, endogenous or exogenous, in a predisposed host.[1] The follicle may serve as a site for cell-mediated immune responses due to enhanced HLA-DR antigen expression, Ɣ-δ T lymphocyte localization within the follicle, and increased Langerhans cell concentration in the infundibular segment of the follicle, germinative sebaceous epithelium, and bulge area.[1] Potential endogenous antigens include heat shock proteins (HSP) and cytokeratin 18 in rheumatoid arthritis patients.[1] Whereas, exogenous stimuli include monosodium glutamate, ginger, upper respiratory infection pathogens, mycobacterium, hepatitis B, and drug therapy in individuals with dysregulated immune systems.[1]

Lesions vary in presentation and appearance as folliculitis, vasculitis, or vesiculopustular or acneiform eruptions preferentially on the trunk, lower extremities, and upper extremities.[1,2] Constitutional symptoms such as fever, arthralgias, or malaise may accompany the lesions.[1] While the histopathological and clinical appearance may suggest bacterial folliculitis, presence of constitutional symptoms should warrant other considerations of systemic diseases.[1] Histopathology reveals neutrophilic or suppurative and granulomatous folliculitis in conjunction with either: 1) perivascular and intramural neutrophilic infiltrate with leukocytoclasia and erythrocyte extravasation, demonstrating a Sweet's-like vascular reaction or 2) fibrinoid necrosis of vessel wall erythrocyte extravasation, demonstrating a leukocytoclastic vasculitis.[1] Tissue cultures and special stains fail to reveal an infectious pathogen.[1]

This case is presented to highlight the clinical presentation of sterile neutrophilic folliculitis. While there is information in the literature regarding its histopathology, there is a paucity of information on its clinical features. The case discussed illustrates a gentleman whose lesions were initially macular then evolved into centrally umbilicated, semi-translucent papules. The lesions then quickly involuted 7 days after initial presentation. Interestingly, the patient continued to have constitutional symptoms and positive blood cultures while the lesions faded.

The finding of sterile neutrophilic folliculitis with vasculopathy does not necessarily indicate the need for an exhaustive systemic disease workup.[1] However, an infectious trigger or underlying systemic illness such as connective tissue disease, inflammatory bowel disease, or Behcet's disease should be considered. Definitive treatment options have not been identified, however management of the underlying condition is the mainstay of treatment at this time.

Conflict of Interest Disclosures: None

Funding: None

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References:

1. Magro CM, Crowson AN. Sterile neutrophilic folliculitis with perifollicular vasculopathy: a distinctive cutaneous reaction pattern reflecting systemic disease. *Journal of Cutaneous Pathology*. 1998;25(4):215-221.
2. Patterson JW. *Weedon's Skin Pathology*. 4th ed. London: Churchill Livingstone; 2016.
3. Patterson JW. *Practical Skin Pathology: a Diagnostic Approach*. Philadelphia, PA: Elsevier/Saunders; 2013.